DEFERVESCENCE KAWASAKI DISEASE ASSOCIATED WITH PARAINFLUENZA TYPE 1 INFECTION

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ABSTRACT

Kawasaki Disease is a systemic vasculitis, associated with acute febrile illness of childhood. It is the leading cause of acquired heart disease in children in developed countries. The causative agent of Kawasaki Disease is still not known [1-2]. Parainfluenza virus causes croup, bronchiolitis, pneumonia, rarely mumps, aseptic meningitis and encephalitis. We report a 4 years 9 month old girl with defervescence Kawasaki Disease associated with Parainfluenza type 1 infection. It is mostly seen in children rarely with adult or infant of less than 2 month age [3].

INTRODUCTION

Kawasaki Disease

Diagnosis of Kawasaki Disease is a tough job to the pediatrician, strong suspicion along with clinical and laboratory data to be conglomerate to diagnosis the disease. The classic Kawasaki Disease was diagnosed if at least 5 of the 6 criteria were met. The diagnosis of incomplete Kawasaki Disease was made in case not fulfilling the criteria for classic Kawasaki Disease but strongly suggested for Kawasaki Disease [4].The patients were classified into the defervesced Kawasaki Disease (dKD) if the symptoms particularly fever resolve without receiving intravenous immunoglobulin (IVIG) or glucocorticoids therapy within 7 days of illness and no fever for more than 3 days [5]. Indolent Kawasaki Disease (iKD) means those of defervesced Kawasaki Disease developed coronary artery lesions (CALS) or required IVIG for ongoing systemic inflammation.

Current Diagnostic criteria for classic Kawasaki Disease

Fever more than 5 days and more than 4 of 5 clinical finding which need not be present at the same time.

1. Extremity changes (Erythema of palm, sole and or edema of hand, feet)
2. Polymorphous exanthema particularly in perineal region with desquamation.
3. Non exudating bulbar limbic spearing bilateral conjunctivitis.
4. Mucosal changes- Erythema of lips , Oropharynx , strawberry tongue, cracked swollen lips.
5. Unilateral cervical lymphadenopathy (›1.5cm in diameter)

Incomplete Kawasaki Disease

If symptoms consistent with Kawasaki Disease along with fever for ≥5 days with 2 or 3 clinical criteria associated with CRP › 30mg/L ESR ≥40 obtained Echo. If more than 3 supplemental lab criteria are present treat for Kawasaki Disease. Supplemental lab criteria are 1) Albumin ≤3gm/dl, 2) platelet count ≥4.5L/µl in 2nd week, 3) white blood corpuscle ≥ 15000 /µl , 4) Urine RE ≥10WBC/HPF(high power field), 5) Anemia for age,6) ALT(alkaline phosphate) raised[6].

Patient characteristics:

Age: 4yr 10 m
Gender: girl
Numbers of days of fever: 7 days
Non suppurative conjunctivitis: +ve
Cervical lymphadenopathy : -ve
Strawberry tongue: +ve
Maculopapular rash : +ve
Pitting edema of hand and toes: +ve
Concomitant viral infection: Para influenza type 1
Echocardiography finding: normal
Z score: normal

Additional findings: ESR 42 , CRP 23.7mg/L, WBC 15800/µl, Hb 10.8gm/dl, platelet 5.8L/ml, liver function test (LFT)-normal.

Perineal rash: Erythematous vaginal mucosa and desquamating rash in groin.

Gastro intestinal track (GIT): Pain abdomen, Ultrasonography of abdomen show sludge within gall bladder, multiple enlarge mesenteric lymph node (LN) with localized omental thickening and loculated collection at ileocecal region.

Kawasaki Disease is the most common systemic vasculitis and 2nd most common cause of pediatric coronary artery Disease. IVIG decrease the coronary artery disease from 20 to 5% [7]. It is seen that some patient experienced spontaneous defervesces within 10 days. They did not received IVIG. The two important reasons of not received IVIG are incomplete presentation and

cost of medicine. Japanese Society of Pediatric Cardiology and Cardiac Surgery 2012 (JSPCCS) also recommend to refrain from IVIG for less severe KD or spontaneous defervesced based on the scoring system [8].

Indication of IVIG
Typical KD: Presence of principal symptoms and who are at risk for coronary artery aneurysm (CAA).
Atypical KD: Not fulfilling the diagnostic criteria and other disease or condition excluded
Less severe or spontaneous defervesced KD: Pediatrician may refrain according to Harada Score.

Role of IVIG in KD
The aim of treatment of KD is to minimize the risk of development of coronary artery lesion (CAL). The arteritis develops by 8 to 9 days. Best result obtained if IVIG be started before day 7 and preferably by before 10 days of illness. Fever resolve within 48 hours of starting IVIG, persisting fever more than 48 hours considered as IVIG resistant KD, requiring second line treatment. Additional IVIG is the most common second line treatment. Other options for second line treatment are intravenous methyl prednisolone pulse (IVMP) 30mg/kg for 3 days with or without an oral glucocorticoid or a single infliximab(IFX) infusion of 5mg/kg.

Complication of KD
Apart from coronary artery lesion(CAL)

Cardiovascular: Myocarditis, Pericardial effusion, Valvular regurgitation, rarely Arrhythmia and cardiac failure.

General: Edema, Hypoalbuminemia, Hyponatremia, Anemia.

Gastro Intestinal Tract (GIT): Cholecystitis, Paralytic ileus, Hepatic dysfunction, Diarrhoea, Vomiting, Dehydration.

Neurological: Altered consciousness, Convulsion.

KD is the leading cause of coronary artery disease in children. Intravenous immunoglobulion (IVIG) is effective to prevent the development of coronary complication but resistance to IVIG may be present. Different scoring system were developed to evaluate the development of coronary artery aneurysm(CAA) and IVIG resistant. The popular scoring system are Harada, Kobayashi, Egami, Formosa, and Sano, Among the five scoring system Harada is significant for predicting CAA risk and Kobayashi for IVIG resistant.

	Harada	kobayashi	Egami
Sensitivity	90%	86%	61-78%
Specificity	51%	68%	81-76%

This scoring system are based on different clinical and laboratory data and tested in different countries with sensitivity and specificity explained by ethnic variability.

The Harada risk score is one most well known risk score and

include 1. WBC count>12000/μl, 2. Hematocrit<35 %, 3. Platelets <350000/μl, 4. CRP>3mg/dl, 5. Albumin<3.5g/dl, 6. Age≤12years , 7. Male sex.

The kobayashi risk score includes 1. Sodium≤133mmol/L-2points, 2. Days of illness at initial treatment≤4days-2points, 3. AST ≥100IU/L-2points, 4. Percentage of Neutrophils ≥80%-2points, 5. CRP ≥10mg/dl-1point, 6. Age≤12months-1point, 7. Platelet count≤300000/cmm-1point.

The Egami risk score includes 1. Age<6months-1point, 2, Days of illness<4-1point, 3. Platelet count<300000/cmm-1point, 4. CRP>8mg/dl-1point, 5. Alanine Amino Transferase >80IU/L-2points.

Incidence of spontaneous defervescence KD
6.9% experienced spontaneous defervescence after diagnosis of KD and were classified in the defervesced KD group [9]. If fever resolve spontaneously 10 days after the onset, IVIG does not need to be administered in the spontaneous defervescence patient provided- 1: normalization of laboratory values and normal ecocardiographic result[7,10] or 2: low severity of the disease.

CAA in spontaneous defervescence disease
It is seen in one study that the incidence of CAA in spontaneous defervescence patient was 5.7% and 4.6% in the treated with IVIG. KD is self limiting but CAA has been reported up to 25% of untreated cases [9].

It was reported in another study the incidence of CAA with defervesence KD is 18.9%that significant higher than treated cases 5.1% [11]. It is address the importance of use of IVIG in the spontaneously defervesced KD patient within 10 days after starting of fever at least in younger patient of <1year of age with Leukocytosis .

Measurement of Coronary Artery Lesion
The main coronary arteries which are considered for measurement- 1: left main coronary artery (LMCA), 2: left anterior descending artery (LAD) and 3: right coronary artery (RCA). The Z score of coronary artery is adjusted by the body surface area using data of Taiwanese children [12].

Definition of coronary artery lesion is a Z score of coronary diameter ≥ +2.5 (13).

Coronary artery aneurysm are sub classified as small (+2.5≤ Z<+5.0), medium (+5.0≤Z<10), and giant (Z≥+10.0) [7].

CONCLUSION
In our patient 4years 9month old girl having defervescent KD associated with para-influenza type-1 infection was admitted at hospital with fever, conjunctivitis , buccal mucosal ulcer , strawberry tongue, vaginal mucosal Erythema, pain abdomen. After spontaneous remission from fever she had periungal desquamation. Echocardiography revealed structurally normal heart, normal coronary arteries. She is under regular follow up and till now no coronary artery abnormality detected though

some study concluded that 18.9% of spontaneously defervesced KD developed coronary aneurysm compared with KD patient who received IVIG within 10 days after fever onset [14].

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